307 Altered body composition in males homozygous for the Delta F508 mutation

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Reduced bone mineral density (BMD) and altered body composition are well recognised in patients with CF. The causes of reduced BMD are multifactorial.

Aim: To assess the effect of genotype and sex on BMD and body composition in adults with CF.

Methods: BMD was measured at the lumbar spine (LS), femoral necks (FN) and total body (TB) by DXA (GE-Lunar Prodigy) and presented as Z scores. Weight, height and BMI were determined at the time of scanning. Height squared indices were derived for fat free mass (FFMI), total body lean (TBLI), total body bone mineral content (TBBMCI) and fat mass (FMI). Patients were divided by sex and categorised as homozygous or heterozygous for the delta F508 mutation.

Results: 246 scans were performed in 125 males, 84 homozygous, mean age 23.8 (SD 5.1); 41 heterozygous, mean age 29.8 (SD 8.5); and 121 females, 76 homozygous, mean age 26.2 (SD 7.1); 45 heterozygous; mean age 26.4 (SD 8.1). There were no significant differences for any parameter for females. Males homozygous for deltaF508 had lower Z LS BMD –1.38 (SD 1.4) compared to heterozygotes –0.65 (SD 1.5) $p = 0.01$, Z TB BMD –0.9 (SD 1.3) compared to 0.02 (SD 1.4) $p < 0.0001$, FFMI 17.9 (SD 2.0) compared to 18.7 (SD 1.9) $p < 0.05$, TBBMCI 0.86 (SD 0.13) compared to 0.96 (SD 0.14) $p < 0.001$ and FMI 3.7 (SD 2.2) compared to 4.7 (SD 2.7) $p < 0.05$

Conclusion: Reduced bone mineral density in CF is multifactorial. Others have described an association with the delta F508 genotype. Our results support the hypothesis that reduced BMD in CF has a genetic component. Unlike other studies we found a difference between homozygous and heterozygous males. This effect was also exerted on FFMI and FMI.

308 Nutritional therapy for catch-up growth in malnourished infants with Cystic Fibrosis – the role of semi-elemental formula with medium chain triglycerides

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Aims: to establish the most efficacious nutritional therapy for catch up growth to the expected growth parameters in infants with cystic fibrosis(CF).

Methods: have been studied 12 infants with CF, diagnosed in our Clinic. All of them were malnourished when the diagnosis of CF was done, with weight for height indexes 50%-85%. Their growth was studied comparing their diets: lactose free formula, semi-elemental formula with hydrolyzed protein and medium chain triglycerides (MCT). Formula was given as bottle feeding and/or nasogastric tube with continuous drip. All the infants received pancreatic enzyme supplementation with Kreon (Solvay) 1000–2500 ui/kg/day.

Results: 7 infants (58%) catch up growth to the normal growth parameters. For 3 infants (25%) the nutritional status improved; 2 infants (17%) remained with the same degree of malnutrition. From the 12 infants, 9 were fed with semi-elemental formula: 6 gained weight and returned to normal growth (66.6%), with formula given for 1 to 6 months, 140–150 kcal/kg/day; 3 infants improved their nutritional status, receiving formula late, after other failing attempts with complete formula or lactose free formula, when they already had severe malnutrition. One infant catch up growth with lactose free formula and solid supplements. 2 infants didn't improve nutritional status: they were fed with lactose free formula, for 110–120 kcal/kg/day and short periods insufficient amounts of semi-elemental formula.

Conclusion: Our study prove the superiority of semi-elemental formula with MCT, associated with pancreatic enzyme supplementation, in the nutritional therapy for malnourished infants with CF.This formula should be administrated early, oral or/and enteral continuous tube feeding, providing 140–150 kcal/kg/day, for 1 month or longer, guided by the severity of malnutrition.

309* Assessment of nutritional status of Italian CF patients

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Aims: To identify trends in growth of CF patients attending tertiary Italian Centers. To explore the relationship between height, weight, and Body Mass Index (BMI) with potential explaining variables.

Methods: Z-score for height, weight, and BMI measured at annual assessment from 2004 to 2005 in patients followed up by 7 CF reference centres were analyzed. The patients were classified in 4 age-groups (g1:0–4 yrs; g2: 5–10 yrs; g3 11–18 yrs; g4 >18 yrs). The effects on growth of Sex, Pancreatic Insufficiency (PI), Meconium Ileus (MI), Respiratory function expressed as % predicted Forced Expiratory Volume in 1 second (FEV1) for sex and age, Cronic Infection by Pseudomonas Aeruginosa (PA), CF Related Diabetes (CFRD), Liver Disease (LD), Gastroesophageal Reflux (GER) were explored.

Results: Data on 981 CF patients (54.5% male) aged 0.30 to 539 months were analyzed. The average growth of Italian CF patients was satisfactory (median >25°pc). There were no gender differences in median Z-score for height, weight, and BMI (males: –0.16, –0.34, –0.02, females: –0.30, –0.42, –0.02, $p = 0.42$; 0.70; 0.65 respectively). Young children and patients close to the age of puberty showed a trend toward lower values of BMI, weight, and height compared with reference. Presence of PI and CF related complications (PA, GER, CFRD, MI, LD) negatively affected height.

Conclusions: Despite the general growth pattern of Italian CF patients is in the normal range, suboptimal nutritional status can occasionally occur, particularly in young children and adolescents.

310 Respiratory muscle strength and nutritional status in Cystic Fibrosis patients

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The aim of this study was to investigate the relationship between respiratory muscle strength and nutritional status in cystic fibrosis (CF) patients.

Thirty-two CF individuals (9–34, 19.98±1.0 years) were divided into: Group A: 12 patients having BMI zscore ≥ 0 and Group B: 20 patients with BMI zscore <0. Nutritional status was determined by BMI (Body weight/heigth^2) BMI zscore, Body Fat %(BFAT%) and Lean Body Mass % (LBM%) measured with bioelectric impetance analysis. Respiratory muscle strength and endurance were calculated from mouth pressures during PEmax, PImax and MVV.

Results were expressed as mean±SEM. Group A: BMI=24.29±0.7, BMI zscore=0.76±0.16,BFAT%=25.43±2.26, LBM%=74.54±2.27, PImax=84.83±6.47, PEmax=113.72±6.0 and MVV=99.83±11.7. PEmax correlated with LBM% ($r = 0.704$, $p = 0.016$) and BFAT% ($r = -0.709$, $p = 0.015$). PI max and MVV did not show significant correlations. Group B: BMI=18.27±0.36, BMI zscore=–1.39±0.19, BFAT%=15.44±1.3, LBM%=85.05±1.3, PImax=99.7±3.22, PEmax=113.5±6.4 and MVV=106.35±13.1. PEmax correlated with LBM% ($r = 0.76$, $p = 0.0001$), BFAT% ($r = -0.67$, $p = 0.002$) and BMI zscore ($r = -0.49$, $p = 0.039$). PI max correlated with LBM% ($r = 0.5$, $p = 0.023$), BFAT% ($r = -0.67$, $p = 0.002$) and BMI zscore ($r = -0.49$, $p = 0.039$). MVV correlated with LBM% ($r = -0.43$, $p = 0.041$), BFAT% ($r = 0.41$, $p = 0.048$) and BMI zscore ($r = 0.45$, $p = 0.03$). The difference in PImax was statistically significant ($t = 2.29$, $p = 0.029$) but there was no significant difference in PEmax and MVV means between the two groups.

Conclusion: Respiratory muscle strength and endurance are related to nutritional status. Up to 25% reduction of BMI does not affect muscle strength and endurance, provided high LBM% is maintained. A decrease in LBM% will first influence muscle strength and then muscle endurance.